Congenital Heart Disease

Balloon Pulmonary Valvuloplasty in Adults with Congenital Valvular Pulmonary Stenosis

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Background: Balloon valvuloplasty has been used for congenital valvular pulmonary stenosis (PS) in pediatric patients as a conventional treatment. The purpose of this study was to further investigate the safety, immediate and long-term results of balloon valvuloplasty in adults with congenital valvular pulmonary stenosis.

Method and Result: We analyzed hemodynamic data of 12 patients who underwent balloon pulmonary valvuloplasty (ages 20-80, mean 41 years). Double-balloon technique was used in 8 patients and single-balloon technique in 4 patients (including 1 case with Inoue balloon). Right ventricle systolic pressure and pulmonary valve peak-to-peak systolic pressure gradient decreased from 101.4 ± 35.0 to 48.8 ± 17.7 mmHg (p < 0.001) and 85.3 ± 37.3 to 29.9 ± 20.0 mmHg (p < 0.001), respectively. Ten patients underwent transthoracic Doppler echocardiography follow-up 2-184 months (mean 68.6 months) after pulmonary balloon valvuloplasty, and peak transvalvular systolic pressure gradient further decreased to 15.1 ± 11.9 mmHg (p = 0.001). All patients had symptomatic improvement, and no major complication or mortality was noted.

Conclusion: Balloon pulmonary valvuloplasty is a safe and effective treatment for adult patients with congenital valvular PS. In our experience, even elderly patients (up to 80 years old) had satisfactory result.

Key Words: Pulmonary stenosis • Balloon valvuloplasty • Adult

INTRODUCTION

Congenital valvular pulmonary stenosis (PS) accounts for most of the etiology of PS, and constitutes about 5 to 10% of all congenital heart disease. The vast majority of patients have thick, conical or dome-shaped pulmonary valve due to fusion of commissures. Occasionally, the valve may be dysplastic, which is related to Noonan’s syndrome. There are various definitions of severity grading, but there is general agreement that transvalvular peak systolic pressure gradient < 25 mmHg is trivial; 25 to 49 mmHg, mild; 50 to 79 mmHg, moderate; ≥ 80 mmHg, severe. Natural history reveals that severe congenital valvular PS has relatively poor long-term prognosis due to right heart failure, especially there is right-to-left shunt.

It is recommended that the indications for intervention should include the following 2 criteria: 1. patients with exertional dyspnea, angina, syncope, or presyncope; 2. asymptomatic patients with normal cardiac output (estimated clinically or determined by catheterization) and transvalvular peak systolic pressure gradient ≥ 30 mmHg. Since 1947 surgical valvulotomy has been performed for critical valvular PS. In 1979, Semb et al. first introduced nonsurgical dilatation of stenotic pulmonary valve by balloon technique in a pediatric patient, and later in 1982, Pepine et al. first described successful balloon valvuloplasty in an adult patient.

During the past 20 years, both short- and long-term

[Additional text not shown]
benefits of this non-surgical procedure in children or infants have been well established. However, the similar data in adults, especially old age patients, are relatively less well-defined. Here we present our experience of balloon valvuloplasty for adult patients with congenital PS.

METHOD

Patient selection
Between 1982 and 2003, balloon pulmonary valvuloplasty (BPV) was performed on 12 patients with congenital valvular PS (4 men and 8 women; age ranged from 20 to 80, mean 41 years). Doppler echocardiography was performed routinely before BPV to evaluate structure heart diseases, and no one had other coexisting congenital heart anomalies except valvular PS. Clinically, all patients were symptomatic, such as with exercise intolerance or effort dyspnea. Their demographic data are shown in the Table 1.

Procedures
Vascular access via femoral vein, right ventricular angiography was done with a Pigtail catheter initially. Hemodynamic data including RV pressure and pulmonary artery (PA) pressure were documented during catheterization with an end-hole catheter. Under electrocardiography and blood pressure monitoring, BPV was performed basically according to the method of Kan et al. and Al-Kasab et al. Briefly, a long exchange guidewire (260 cm) was used to advance the balloon to the pulmonary valve site. Single-balloon technique was performed in 4 patients (including 1 case with Inoue balloon) via unilateral femoral vein, with the balloon sized about 1.3 times the annulus diameter. Double-balloon technique was performed in 8 patients via bilateral femoral vein with each balloon sized about 0.65 times the annulus diameter. Successful dilatation was indicated by the disappearance of the waist around the balloon under cineangiography (Figure 1B and C). Usually, repeated balloon dilatation 2-3 times was performed and each inflation time was no more than 5 seconds. After balloon dilatation, the hemodynamic data were checked and RV angiography was done again for final result. Patients had OPD follow-up with prescription of beta-blocker, and Doppler echocardiography at various time intervals was arranged to obtain pulmonary valve pressure gradients.

Table 1. Hemodynamic data before and after balloon pulmonary valvuloplasty and Doppler pressure gradient measurement at follow-up

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age/gender</th>
<th>Balloon</th>
<th>RVP (mmHg)</th>
<th>PAP (mmHg)</th>
<th>PG (RV-PA) (mmHg)</th>
<th>Follow-up (Months)</th>
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<td>Post-</td>
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<td>Post-</td>
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<td>37</td>
<td>9</td>
<td>15</td>
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<tr>
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<tr>
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</table>

P values compare differences before and after BPV. *P value compares difference after BPV and at follow-up.

Abbreviations: RVP = right ventricle pressure; PAP = pulmonary artery pressure; PG (RV-PA) = pressure gradient between right ventricle systolic pressure and pulmonary systolic pressure.
**Statistics**

All measured hemodynamic values are expressed as mean ± SD (standard deviation). Paired Student’s t test was used to compare data differences, and $p$ value < 0.05 was considered to be significant.

**RESULTS**

All patients had typically “dome-shaped” pulmonary valve, post-stenotic dilatation of the main PA and marked trabeculation of RV on right ventricular angiography. Morphology also revealed a rapid jet across the stenotic valve (Figure 1A). After BPV, the transvalvular flow became much wider because the opening of valve was no longer narrowed (Figure 1D).

After BPV, RV systolic pressure decreased from $101.4 \pm 35.0$ to $48.8 \pm 17.7$ mmHg ($p < 0.001$); RV-to-PA peak systolic pressure gradient decreased from $85.3 \pm 37.3$ to $29.9 \pm 20.0$ mmHg ($p < 0.001$) (Figure 2A and B). There was also a significant change of PA systolic pressure from $16.0 \pm 5.7$ to $19.7 \pm 6.9$ mmHg ($p = 0.025$) (Table 1). Transvalvular pressure gradient at repeated follow-up by precordial Doppler echocardiography were obtained in 10 patients at a mean interval of 68.6 months (ranged from 2 to 184 months) after the initial BPV, and further drop to $15.1 \pm 11.9$ mmHg ($p = 0.001$) was demonstrated (Figure 2B).

All patients had symptomatic improvement. Although systemic hypotension (below 100 mmHg) and transient episodes of arrhythmia occurred in some cases during balloon dilatation (especially with the use of single-balloon technique), there was no major complication, such as severe pulmonary regurgitation (PR), tamponade or mortality, ever noted. No patients required post-catheterization care in ICU or any emergent surgical management. All patients were discharged within 1 week.

![Figure 1](image_url)

*Figure 1. An adult patient with congenital valvular pulmonary stenosis. Before BPV, RV angiography (anteroposterior view) showed dome-shaped of pulmonary valve (arrow), rapid jet across the stenotic valve, and post-stenotic dilatation of main PA (asterisk) (A); During BPV, the double-balloon technique was used. There was full dilatation over the pulmonary valve until loss of the waist of balloons (B, C); After BPV, the transvalvular flow became much wider, which meant the valve opening was no longer narrowed (D).*
DISCUSSION

Our results demonstrate that BPV is a safe and effective procedure in treating adult patients with congenital valvular PS.

BPV has become the choice of treatment for valvular PS since the first series reported by Kan et al. in 1982, and has almost replaced surgical valvotomy in pediatric patients. The double-balloon technique was first reported by Al-Kasab et al. in 1987. The use of 2 balloons may permit a small amount of blood flow between them even during full dilatation, and leads to fewer hemodynamic changes. Another technique, the use of Inoue balloon, which was first reported by Lau et al. in 1993, also has advantages over the single-balloon technique because it is size-adjustable, making stepwise dilatation possible, and due to its short and self-positioning characters, minimizing the possible injury to RV infundibulum or main PA. But Inoue balloon has disadvantages including necessity of a large sheath, rigid property and costly expense. Consequently, we favor the double-balloon technique now.

According to previous studies, the independent predictors of long-term result after BPV in pediatric patients are: 1. valve morphology; 2. ratio of balloon to annulus diameter; and 3. immediate post-dilatation pressure gradient of the pulmonary valve. Poor long-term result is observed if the valve is dysplastic or the ratio of balloon to annulus diameter < 1.2 or residual transvalvular pressure gradient > 36 mmHg. However, some authors have claimed that it is not necessary to use a larger balloon (ratio of balloon-to-annulus diameter > 1.2) in adults as in children because adults have a much lower restenosis rate than children (4.8% vs. 19%) and there is no clear relationship between balloon size and hemodynamic results. Most authors suggested that balloon to annulus ratio should not exceed 1.5 due to the higher risk of severe PR or annular laceration, unless there is a residual RV to PA pressure gradient greater than 36 mmHg.

In the literature, the degree of PR was higher in most studies after BPV. The incidence of newly developed PR is 13 to 39%, but most cases are only mild, and less frequent than following surgical valvotomy, which is about 60%. Even moderate to severe PR is able to be tolerated and does not influence the clinical outcome. Major complications of BPV have been reported in pediatric patients, including death (0.2%) and cardiac perforation (0.1%). Among adult patients, 1 death was reported by Hermann et al., and 1 case of cardiac tamponade by Kaul et al. In contrast, surgical valvotomy has a higher mortality rate of 1.5 to 2%. In our series, we did not encounter those complications mentioned above. Only mild hypotension and transient episodes of arrhythmia were noted during the procedures which did not necessitate terminating the procedure. Besides, 3 patients, aged of 68, 71 and 80 years old, could tolerate the whole treatment course as well with good results.

Generally speaking, after BPV, there is a decrease of
RV systolic pressure ranged from 39 to 71%, and a decrease of transvalvular pressure gradient ranged from 45 to 93%\(^{13,18,19,22,23,27,30,34,36,37}\). In our study, the results were equal to those of other centers: 52% reduction of RV pressure and 64% reduction of transvalvular pressure gradient on average. Significant infundibular PS is a problem which may cause high residual pressure gradient after BPV. It was suggested by experts that myomectomy should be performed if immediate post-procedure RV pressure still exceeds 100 mmHg.\(^{38}\) Reflex tachycardia and improvement of contraction would cause an increase of cardiac output, but also worsen the obstruction of RV outflow tract and raise its pressure gradient.\(^{39}\) Cases of infundibular spasm after BPV were reported by Al-Kasab et al.\(^{18}\) However, among our patients, we did not find such conditions occurring. In addition, RV infundibular hypertrophy secondary to PS usually regresses gradually after the procedure of BPV, which may be accelerated by the administration of oral beta-blockers. Therefore it results in sustained improvement of transvalvular pressure gradient.\(^{40}\) Like other studies,\(^{27,34,37}\) our data also show similar pattern of improvement of transvalvular pressure gradient at long-term follow-up (Figure 2B).

In conclusion, BPV is a safe, effective and reliable treatment for adult patients with congenital valvar PS. The immediate and long-term results are excellent. Even in elderly patients, BPV is also a suitable treatment modality.

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REFERENCES

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成人先天性肺動脈瓣狹窄以氣球擴張術治療之效果

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台北市  台北市立仁愛醫院  設內科
台北市  台大醫院  心臟內科

背景 先天性肺動脈瓣狹窄以氣球擴張術 (心臟瓣膜成型術) 治療在小兒科已經行之有年，本研究主要評估成人先天性肺動脈瓣狹窄以氣球擴張術治療之效果。

方法與結果 經統計 12 位接受氣球擴張術治療之成年病患 (平均 41 歲，其中 3 人高齡達 68、71 及 80 歲)，8 例為雙氣球、4 例為單氣球擴張術 (其中 1 例使用 Inoue 氣球)，分析其術前、術後及長期追蹤之血液動力學之各項參數。我們發現術後右心室收縮壓立即由 101.4 ± 35.0 毫米汞柱減少為 48.8 ± 17.7 毫米汞柱，肺動脈瓣壓力差立即由 85.3 ± 37.3 毫米汞柱減少為 29.9 ± 20.0 毫米汞柱。十位病患以超音波追蹤 (平均 68.6 個月)，肺動脈瓣壓力差逐漸減少為 15.1 ± 11.9 毫米汞柱。全部病患症狀皆獲得改善，而且沒有出現併發症。

結論 以氣球擴張術治療成人先天性肺動脈瓣狹窄，是十分安全又有效的治療方式，甚至施行於高齡老人也相當成功。

關鍵詞：肺動脈瓣狹窄、氣球擴張術、成人。